A Retrospective Analyses of Newborns Operated Due to Midline Spinal Closure Defect: A Single Center Experience

Spinal Orta Hat Kapanma Defekti Nedeniyle Opere Edilen Yenidoğanların Retrospektif İncelenmesi: Tek Merkez Deneyimi

Spinal Midline Closure Defects

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Öz

Amaç: Bu çalışmada spinal orta hat kapanma defekti nedeniyle opere edilen yenidoğanların retrospektif incelenmesi amaçlanmıştır. Gereç ve Yöntem: Sunulan çalışmada 2007-2014 yılları arasında kliniğimizce opere edilen 62 yenidoğan değerlendirilmeye alındı. Görüntüleme yöntemleriyle eşlik eden kranial ve spinal patolojiler incelendi. Yapılan operasyonun zamanlaması (ilk 3 gün, 3 gün sonrası) ve yöntemi (primer, sekonder) belirlendi. Ek olarak hidrosefali gelişimi takip edildi, hidrosefali nedeniyle şant takılan hastaların takip sürecinde şant enfeksiyonu oranı da belirlendi. Bulgular: Çalışma popülasyonundaki 62 yenidoğanın 38'i (%61.2) kız, 24'ü (%38.8) erkekti. Ortalama takip süresi 25±6.9 aydı. Orta hat defekti hastaların 1'inde servikal bölgede (%1.6), 7 sinde torakal bölgede (%11.9), 54'ünde lumbosakral bölgede (%87.1) idi. Hastaların 54'ü (%87.1) meningomylosel, 8'i (%12.9) meningosel nedeniyle opere edilmişti. Meningosellilerin 1'ine, meningomyelosellerin 48'ine hidrosefali nedeniyle şant takıldı. Cilt defektlerinin 9'u sekonder (%14.5), 53'ü primer kapatılmıştı (%85.5). Yenidoğanların 21'i doğumdan sonraki ilk 3 gün içerisinde 41'i ilk 3 günden sonra opere edilmişti. 13 hastada (%21) şant enfeksiyonu saptandı. Tartışma: Orta hat kapanma defektine intrakranial en sık eşlik eden patoloji hidrosefali, spinal en sık eşlik eden patoloji syringomyeli ve tethered korddu. Doğumdan sonra ilk 3 gün içerisinde opere edilen yenidoğanlarda şant enfeksiyonu oranı daha düşüktü. Defektin primer yada sekonder kapatılması şant enfeksiyon oranını etkilememişti.

Anahtar Kelimeler

Orta Hat Spinal Kapanma Defekti; Hidrosefali; Yenidoğan

Abstract

Aim: This study aimed to retrospectively investigate the newborns who were operated due to midline spinal closure defect. Material and Method: 62 newborns who were operated in our clinic between 2007-2014 were evaluated in this study. Accompanying cranial and spinal pathologies were examined by imaging methods. The timing (during first 3 days, after first 3 days) and the method (primary, secondary) of the surgery were determined. In addition, the shunt infection rate was also evaluated in the follow-up period of the patients who underwent VP shunt placement due to hydrocephalus. Results: A total of 62 newborns were included in the study; 38 (61.2%) were female and 24 (38.8%) were male. The mean follow-up time was 25±6.9 months. The midline defect was localized in the cervical (n:1, 1.6%), thoracal (n:7, 11.9%), and lumbosacral regions (n:54, 87.1%). 21 of the newborns were operated within the first 3 days after birth and 41 of the newborns were operated after the first 3 days. 8 (12.9%) patients were operated due to meningocele and 54 (87.1%) patients were operated due to myelomeningocele. 1 of 8 patients with meningocele and 48 of 54 patients with myelomeningocele underwent VP shunt placement due to hydrocephalus. 9 (14.5%) skin defects were closed secondarily and 53 (85.5%) of skin defects were closed primarily. Shunt infection was detected in 13 (21%) patients. Discussion: The most common intracranial pathology accompanying midline closure defect was hydrocephalus and the most common spinal pathologies accompanying midline closure defect were syringomyelia and tethered cord. Shunt infection rate was lower in the newborns who were operated within the first 3 days after birth. Primary or secondary closure of skin defect did not affect the shunt infection rate.

Keywords

Spinal Dysraphism; Hydrocephalus; Newborn

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Introduction

Midline closure defects is a general name for a group of anomalies related to neural tissues, dura mater and to the surrounding mesenchymal structures (muscle, skin, bone) that arise due to the failure of neural fold closure. This developmental disorder is related to spinal cord, vertebrae, skin, and muscles. Most are in the midline and dorsal region. Although it can be seen in any region of the spinal cord, it is most commonly seen in the lumbosacral region. Meningomyelocele develops when the spinal nerves or nerve roots protrude outward from the skin and bone through a large defect. Meningocele develops when the thecal sac expands from the region where the spine is defective and does not contain nerve tissue.

Midline closure defects are the second most common cause of neuromotor dysfunction in children, second to cerebral palsy [1]. There is not enough data in the literature about the follow-up and treatment of midline closure defects in Turkey [2]. In this study, it was aimed to retrospectively investigate the patients who were operated due to meningocele and myelomeningocele in Eskişehir Osmangazi University between 2007-2014 and to contribute to the literature.

Material and Method

This study included 62 newborns who were operated in our clinic due to a midline closure defect within the first month of their lives. 13 patients were excluded from the study (very low birth weight-under 1500 grams, n:3, applied to our center more than one month after birth, n:3, did not complete follow up-up period, n:7)

All newborns were assessed neurologically in the preoperative and postoperative period. In the preoperative period, brain computed tomography (CT) was performed in all patients and spinal magnetic resonance imaging (MRI) was performed in 26 patients. The purpose of surgical treatment is to repair a structurally damaged sac, to prevent central nervous system infection by creating a barrier between the external environment and the spinal canal, and to provide a suitable environment for cerebrospinal fluid (CSF) circulation. The sac repair was performed by microsurgical method. The neural plate was first separated from the surrounding arachnoid membrane-ectoderm elements and a neural tube was reconstructed. The border between dura and dermis was determined. Then, dura was cut around the dermis border, separated from the subcutaneous tissue, and mobilized towards the midline. It was closed in a watertight fashion with 4-0 silk. The skin was dissected from the lateral fascia and small defects were also closed primarily. The wide skin defects were closed secondarily with a flap or a graft taken from the leg.

A ventriculo peritoneal (VP) shunt was inserted in the same session for those who had hydrocephalus before the sac repair operation. For those who had hydrocephalus in the follow-up period, the VP shunt was inserted at the time of detection. Indications for inserting a VP shunt were determined as unexplained apnea and bradycardia, the presence of ventriculomegaly in imaging methods (developing rapidly and progressively), CSF leak in the area in which the sac is closed, and Chiari Type II malformation with hydrocephalus [3-5].

The SPSS 21.0 software program was used for statistical analysis. Values were given as mean ± standard deviation. The Chi-square and Bonferroni tests were used for comparison of the data. Significance level was considered as $p < 0.05$.

Results

Of 62 newborns in the study, 38 (61.2%) were female and 24 (38.8%) were male. The mean follow-up time was 25±6.9 months. Of the patients with a midline defect, 1 (1.6%) was in the cervical region, 7 (11.9%) were in the thoracic region, and 54 (87.1%) were in the lumbosacral region. 8 (12.9%) patients were operated due to meningocele and 54 (87.1%) patients were operated due to myelomeningocele (Table 1).

Table 1. Demographic characteristics of the patients

	Meningocele, n:8	Myelomeningocele, n:54
Gender, n	Female:4, Male:4	Female:34, Male:20
Location, n	Cervical:1	Cervical: -
	Thoracic:1	Thoracic:6
	Lumbosacral:6	Lumbosacral:48
Neurological condition, n	No deficit:7	Paraparetic:1
	Paraparetic:11	Paraplegic:43
Hydrocephalus VP shunt, n	1	48
Shunt infection, n	-	13
Sac repair, n	First 3 days: -	First 3 days: 21
	After : 8	After: 33
Skin defect, n	Primary: 8	Primary: 45
	Secondary: -	Secondary: 9

21 of the newborns were operated within the first 3 days after birth and 41 of the newborns were operated after the first 3 days. In the follow-up period, shunt infection developed in 1 (4.7%) of the 21 patients who were operated within the first 3 days after birth and in 12 (29.2%) of 41 patients who were operated after the first 3 days. There was a statistically significant difference between the two groups in terms of shunt infection rate ($p<0.001$), (Table 2). 9 (14.5%) of the skin defects were closed secondarily and 53 (85.5%) of the skin defects were closed primarily. In the follow-up period, shunt infection developed in 2 (22.2%) of the patients with skin defects that were closed secondarily and in 11 (20.7%) of the patients with skin defects that were closed primarily. There were no statistically significant differences between the two groups in terms of shunt infection rate ($p>0.05$) (Table 2). 1 of 8 patients with meningocele and 48 of 54 patients with myelomeningocele underwent VP shunt placement due to hydrocephalus. 15 of these underwent VP shunt placement simultaneously with the sac repair operation and 33 of these underwent VP shunt placement in the follow-up period. Shunt infection developed in 4 (26.6%) of the patients who underwent VP shunt placement simultaneously with the sac repair operation and in 9 (27.2%) of the patients who underwent VP shunt placement in the follow-up period. There were no statistically significant differences between the two groups in terms of shunt infection rate ($p>0.05$), (Table 2).

In the preoperative period, brain CT was performed in all patients and spinal MRI was performed in 26 patients. The most

Table 2. Shunt infection rate according to time of the sac repair operation, technique of closure of skin defect, and whether VP shunt was placed simultaneously with the sac repair operation.

	Shunt infection, n, (%)
Operation within the first 3 days, n:21	1 (4.7)
Operation after the first 3 days, n:41	12 (29.2)
p	<0.001
Primary, n:53	11 (20.7)
Secondary, n:9	2 (22.2)
p	>0.05
VP shunt placement simultaneous with the sac repair n:15	4 (26.6)
VP shunt placement in the follow-up period, n:33	9 (27.2)
p	>0.05

common intracranial pathologies accompanying midline closure defect were hydrocephalus, chiari malformation, colpocephaly, and dysgenesis of the corpus callosum, respectively (Table 3). Of the newborns who underwent spinal MRI, 3 had meningocele and 23 had meningomyelocele. The most common spinal

Table 3. Intracranial pathologies accompanying midline closure defect. (*The patients who developed hydrocephalus during the follow-up period were also included).

	Meningomyelocele, n:54	Meningocele, n:8
Hydrocephalus *, n	48	1
Chiari malformation, n	26	1
Dysgenesis of the corpus callosum, n	11	1
Colpocephaly, n	16	-
Neuroglial cyst, n	1	-
Neuroepithelial cyst, n	2	-
Craniosynostosis, n	-	1
Holoprosencephaly, n	1	-
Septo-optic dysplasia, n	1	-
Cephalocel, n	-	1
Periventricular leukomalacia, n	1	-
Subdural hygroma, n	1	-

Table 4: Spinal pathologies accompanying midline closure defect.

	Meningomyelocele, n:23	Meningocele, n:3
Tethered cord, n	7	-
Diastematomyelia, n	3	1
Scoliosis, n	5	2
Syringomyelia, n	7	-
Dermal sinus, n	3	1
Intradural lipoma, n	3	-
Hemivertebra, n	2	-
Terminal ventricle, n	1	-
Sacral agenesis, n	1	-

pathologies accompanying midline closure defect were syringomyelia, tethered cord, and scoliosis (Table 4).

Discussion

Spinal dysraphism is the general name for congenital anomalies affecting dorsal midline structures (the skin, muscles, spinal column, neural tube, and structures surrounding the neural tube). If it is noticeable upon the skin, it is called spina bifida aperta (for example, meningocele and meningomyelocele). If the overlying skin is normal, it is called spina bifida occulta (for example, diastematomyelia, tethered cord, and intradural lipoma) [6-7]. Prognosis is poor especially in advanced forms (for example, myeloschisis and meningomyelocele type 1) of spina bifida aperta. In the past, many clinicians have argued that treatment for meningomyelocele was unnecessary, but the survival time of these infants has now been prolonged with treatment, especially with the multidisciplinary approach used today [8-9]. Sequel-free recovery has become possible with microneurosurgical methods in non-advanced forms such as meningocele.

In this study, 62 patients who were operated in our clinic due to spina bifida aperta during the neonatal period were retrospectively investigated. The shunt infection rate was found to be significantly lower in the newborns who were operated within the first 3 days after birth than the newborns who were operated after the first 3 days. These results are consistent with the literature [10-12]. After the first three days, bacterial pathogens start to colonize and create a favorable environment for infection. Early surgery has some advantages such as reducing exposure of the spinal cord to the external environment, preventing hydroelectrolytic imbalance due to CSF leak, and eliminating the probability of sac rupture [13]. Moreover, there are some studies reporting that early surgery reduces the length of hospital stay and affects cognitive functions positively and decreases the incidence of urinary incontinence [14].

In our study, the most common intracranial pathology accompanying midline closure defect was hydrocephalus. 15 (27.7%) of 54 patients with meningomyelocele had hydrocephalus that required a shunt at first examination and the VP shunt was placed simultaneously with the sac repair operation. In the follow-up period, 33 patients also developed hydrocephalus and 48 (88.8%) of 54 patients with myelomeningocele underwent VP shunt placement. In the literature, the need for a shunt in children with myelomeningocele has been reported as 80-85% [15-16]. In our study, all 6 patients with meningomyelocele in the thoracic region and 42 (87%) of 48 patients with meningomyelocele in the lumbosacral region underwent VP shunt placement. It has also been reported in the literature that the need for a shunt was higher in those with meningomyelocele in the thoracic region compared to those with meningomyelocele in the lumbosacral region [13].

26 of 62 patients were preoperatively screened with spinal MRI; syringomyelia and tethered cord were detected in 7 (27%) patients. It has also been reported in the literature that the combination of meningomyelocele-syringomyelia was 50-80% [17-18] and the combination of meningomyelocele-tethered cord was 30% [19]. The low rates in our study can be explained by the fact that MRIs were performed during the first month. Are the concomitant anomalies such as scoliosis, tethered cord, and syringomyelia separate congenital pathologies or a result of the closure defect? Despite its limitations (inadequate imaging data in the follow-up period), this study suggests that the answer to both questions might be yes, that the anomalies could be separate pathologies or the result of a closure defect."

Primary or secondary closure of skin defect did not have any effect on shunt infection. The shunt infection rate was similar in both groups. In the literature, there is no study showing a re-

lationship between primary and secondary closure method and shunt infection. However, the length of hospital stay, duration of operation, and intraoperative blood loss were higher in the patients with skin defects that were closed secondarily [20-21].

Conclusion

In our study, the most common intracranial pathology accompanying midline closure defect was hydrocephalus and the most common spinal pathologies accompanying midline closure defect were syringomyelia and tethered cord. Shunt infection rate was lower in the newborns who were operated within the first 3 days after birth. Primary or secondary closure of skin defect did not have any effect on shunt infection.

Competing interests

The authors declare that they have no competing interests.

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